

Wunderlich syndrome following rupture of a renal angiomyolipoma

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Wunderlich syndrome refers to spontaneous non-traumatic renal bleeding into the subcapsular and/or perirenal space. The appropriate treatment for this condition depends on the diagnosis of perinephric hemorrhage and the determination of its cause. Radiology therefore plays an important role in the evaluation of patients with this clinical problem. We present the case of a lady who had spontaneous perinephric hemorrhage following rupture of a renal angiomyolipoma (AML) along with a short review of this syndrome.

Case

A 40-year-old, previously healthy woman was brought to our emergency department in a state of shock following sudden severe left flank pain. On examination, the patient was drowsy, pale, cold and sweating, with a pulse of 120 and no recordable blood pressure. She had severe tenderness in the left flank and resisted further examination. The patient was rapidly resuscitated with IV fluids. Her blood hemoglobin and hematocrit levels, which were 9.7 g/dL and 0.29, respectively, dropped to 6.8 g/dL and 0.19 about 6 hours later. The coagulation profile was normal. A urine test for pregnancy was negative. An ultrasound examination of the abdomen showed a large retroperitoneal lesion of heterogeneous echogenicity displacing the left kidney medially. There was no pelvic mass. A provisional diagnosis of retroperitoneal mass/hemorrhage was made and a CT of the abdomen was deemed necessary for further evaluation.

The CT of the abdomen done soon after showed a large left perinephric hematoma displacing the left kidney medially (Figure 1). A hypodense mass of fat density without calcifications was seen in the inter-polar region of the left kidney, poorly margined from the peri-

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Figure 1. CT of the renal region prior to administration of IV contrast shows a heterogeneous attenuation perinephric hematoma displacing the left kidney.

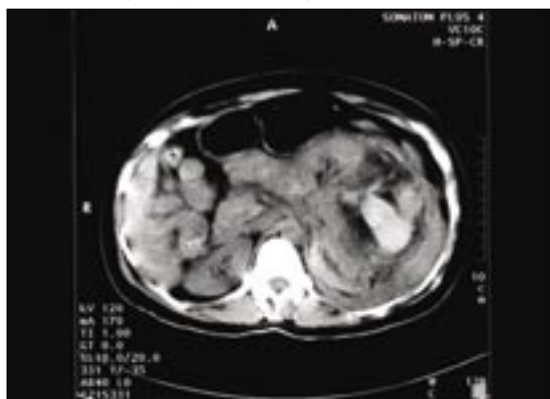
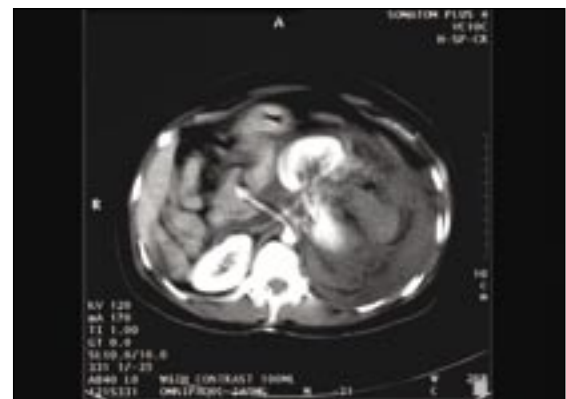


Figure 2. Contrast-enhanced CT shows a mass with fat attenuation [thin arrow: -31 HU] in the left renal cortex.



nephric hematoma (Figure 2). The CT findings were thus consistent with perinephric hematoma secondary to rupture of a renal AML. The rest of the abdomen was normal. The patient received multiple blood transfusions, which corrected her hemoglobin level. She had no further hemorrhage. The patient later underwent left partial nephrectomy and the renal mass was confirmed to be an AML by histopathology. Follow-up ultrasounds at 3 and 6 months post surgery found no new renal lesions.

Discussion

The clinical picture of spontaneous renal bleeding confined to the subcapsular and perinephric space in patients with no known underlying cause was first described by Wunderlich in 1856.¹ Patients may present with the classic triad of symptoms—acute lumbo-abdominal pain, a palpable mass and general deterioration as hypovolemic shock.² Causes described for Wunderlich syndrome include benign and malignant renal tumors, vascular lesions like polyarteritis nodosa, renal infections, nephritis, previously undiagnosed hematological conditions and anatomical lesions like cysts and hydronephrosis.² The commonest cause of spontaneous renal hemorrhage in most series is an AML.³ In our patient the cause for spontaneous perinephric hemorrhage was renal AML. It must be noted that the Wunderlich syndrome is different from Herlyn-Werner-Wunderlich syndrome, which is a malformation characterized by an ipsilateral blind vagina, Gartner duct cyst and ipsilateral renal agenesis.⁴

AML is a choristoma (benign tumor composed of tissues not normally occurring within the organ of origin) made of fat, smooth muscle and thick-walled blood vessels. The abundant abnormal elastin-poor vascular structures in AML make these lesions prone to aneurysm formation and rupture.⁵ Eighty percent of AMLs are sporadic, occurring usually in middle-aged women while the remaining 20% are associated with tuberous sclerosis.⁶ Sporadic AMLs are generally solitary and unilateral, occurring more on the right side. AMLs that occur in association with tuberous sclerosis on the other hand, manifest at a younger age, are likely to be larger and bilateral, and are prone to grow and need surgical treatment.⁷ Fifteen percent of patients with renal AML present with hemorrhage.⁸ Our patient had no evidence of tuberous sclerosis.

The appropriate treatment for patients with spontaneous perinephric hemorrhage depends on the diagnosis that a hemorrhage has occurred and

on the determination of its cause.⁹ Radiology has an important role in addressing these issues. CT is the method of choice for the demonstration of perirenal hemorrhage, with a sensitivity of 100%.³ However, a CT performed at the time of hemorrhage is only moderately successful in identifying the renal neoplasm causing hemorrhage, with a reported sensitivity being 0.57.³ However, it has been found to identify all cases of Wunderlich's syndrome due to AML.² A confident diagnosis of AML can be made by CT by demonstrating the fat content of these lesions.^{2,3,8} Other renal tumors like renal cell carcinoma, liposarcoma, myolipoma, lipoma, oncocytoma, and Wilm's tumor may also show fat content. However, it is felt that a renal cortical mass showing predominantly fat attenuation of less than -20 HU can be diagnosed as an AML, particularly if there is no or little calcification in the lesion.^{10,11} If the initial CT study shows no mass responsible for the hemorrhage, it has been suggested that angiography should be done as it can reveal vascular lesions not shown by CT and also facilitates embolisation.¹²⁻¹⁴ In cases where angiography fails to identify a cause for spontaneous perinephric hemorrhage, a repeat CT has been recommended to detect the causative lesion in patients who can be stabilized medically, as resorption of the hematoma permits detection of a tumor previously hidden by blood.^{9,13} Ultrasonography has been found to be only moderately useful in identifying renal hemorrhage and in differentiating the renal mass and clotted blood.^{2,3,8,15} Biopsy is only rarely useful in the diagnosis of renal AML.⁸

Once a patient is diagnosed with spontaneous perinephric hemorrhage due to AML, the treatment options are either surgery or therapeutic embolisation. Embolisation is extremely useful in the setting of acute hemorrhage due to rupture of renal AML.^{2,8} The benign nature of AML supports a partial nephrectomy or other nephron sparing surgery.⁸ Surgery also facilitates a pathological diagnosis. However, it is felt that if the patient can be stabilized medically during the acute phase of spontaneous perinephric hemorrhage, a nephrectomy can be deferred.^{7,13,15} In a review of the diagnosis and management of 7 cases of Wunderlich syndrome, Cubillana et al also found conservative management to be the most acceptable option, unless a malignant pathology could be demonstrated.¹⁶

In conclusion, we have presented a case of Wunderlich syndrome due to rupture of a renal AML, which is the commonest cause reported in most series. CT plays an important role in the man-

agement of patients presenting with this syndrome, but as illustrated in our patient, by revealing the underlying cause as well.

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